Isolated Single Coronary Artery Presenting as Acute Coronary Syndrome: Case Report and Review

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Abstract

Congenital single coronary artery is commonly associated with complex congenital heart diseases and manifests in infancy or childhood. But isolated single coronary artery is a rare congenital anomaly which can present as acute coronary syndrome in adults. The aim of the work is to discuss on isolated single coronary artery in two adults presenting as acute coronary syndrome. The first case underwent coronary angiography (CAG) through right radial route, but switched over to femoral for confirmation of diagnosis and due to radial spasm. An aortic root angiogram was done to rule out presence of any other coronary ostia. It revealed a single coronary artery originating from right sinus of valsalva. After giving rise to posterior descending artery branch at crux, it continued in the atrioventricular groove to the anterior basal surface of the heart and traversed as anterior descending artery. There was no atheromatous occlusive stenosis. This is R-I type single coronary artery as per Lipton classification. In the second case, angiography was completed through right radial route. It revealed a single coronary artery arising from right aortic sinus. Anterior descending and circumflex branch were originating from proximal common trunk of the single coronary artery and supplying the left side of the heart. The right coronary artery has diffuse atheromatous disease without significant stenosis in any major branch. This is R-III C type as per Lipton classification. A coronary anomaly of both origin and course is very rare. It may be encountered in adults evaluated for atherosclerotic coronary heart disease. Knowledge and understanding of anatomical types of this congenital anomaly will reduce time, anxiety, complications during CAG and cardiac surgery.

Keywords

- single coronary artery
- coronary angiography
- multi-slice computerized tomography coronary angiography
- Lipton classification

Isolated single coronary artery is a rare congenital anomaly where only one coronary artery arises from the aortic trunk by a single coronary ostium and supplies the entire heart, regardless of its distribution.[1] As an isolated finding the occurrence is very low (0.024–0.044%).[2] However, in association with certain other congenital anomalies, like persistent truncus arteriosus, tetralogy of fallot, and pulmonary atresia, it is found considerably more frequently in some series with 17 to 18.5%.[3–5] According to Desmet et al,[6] the first case of single coronary artery was reported by Thebesius[7] in 1716. Until 1950, no more than 45 cases of single coronary artery had been published, all discovered at autopsy. In 1967, the first antemortem diagnosis was made by coronary angiography (CAG).[8] In 1979, Lipton et al[9] proposed a very useful angiographic classification, further modified in 1990 by Yamanaka and Hobbs.[2]

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Since most patients with a single coronary artery are asymptomatic, diagnosis is usually an incidental finding on noninvasive imaging. In symptomatic patients invasive CAG clinches the diagnosis. In this study report, selective coronary arteriography was performed in both the patients and recorded with cineradiography.

Case Details

Our first case was an elderly (72 years) male, diabetic, hypertensive and presented with acute onset retrosternal chest pain, sweating, and breathlessness. On admission his blood pressure (BP) was 150/100 mm Hg, and heart rate was regular with 60 beats per minute. Electrocardiogram (ECG) showed remarkable ST-T wave depression in lead I, aVL, V4-V6. Echocardiogram of the heart showed no regional wall motion abnormality with good left ventricular systolic function. The cardiac troponin-I level was elevated (0.9 ng/mL). Lipid profile revealed high total cholesterol (240 mg/dL), high low-density lipoprotein (LDL) cholesterol (160 mg/dL), and low normal high-density lipoprotein (HDL) cholesterol (40 mg/dL). Blood sugar value was 240 mg/dL. The patient was managed with double antiplatelet agent (aspirin 150 mg and clopidogrel 75 mg once daily), low-molecular-weight heparin, β blocker, angiotensin converting enzyme (ACE) inhibitor, statin, nitrate, and insulin for diabetes. His CAG showed a single coronary artery originating from right sinus of valsalva, traversed in its normal course. After giving rise to PDA branch at crux, it continued in the AV groove to the anterior basal surface of the heart and traversed as anterior descending artery as its terminal branch, as shown in ►**Figs. 1** and **2**. There was no significant atheromatous occlusive stenosis. This is a case of isolated single coronary artery R-I type as per Lipton scheme. CAG was started through right radial route, but switched over to right femoral route as it was not sure regarding the presence or absence of left coronary ostium and also the patient developed radial spasm due to increase in procedural time. An aortic root angiogram, via 6 French pigtail catheter, was done to rule out the presence of any other coronary ostia. The patient's relatives were explained about the congenital isolated single coronary artery syndrome and need for coronary artery bypass surgery (CABG). They did not agree for CABG; hence, the patient was discharged in stable condition on optimal medical management including double antiplatelet (aspirin 150 mg, and clopidogrel 75 mg once daily), β blocker (metoprolol succinate), ACE inhibitor (ramipril), statin (rosuvastatin) and oral antidiabetic agent (glimepiride + metformin).

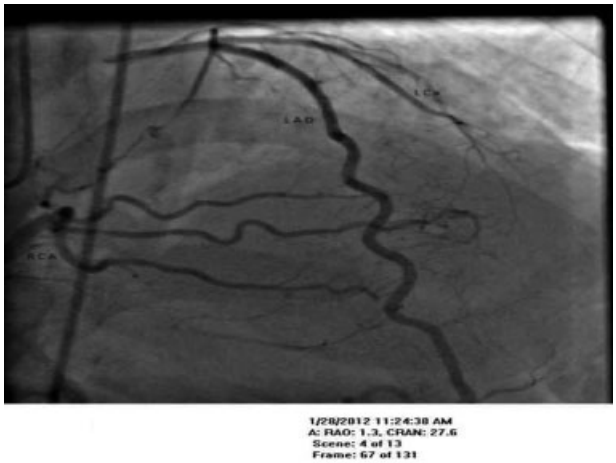

Fig. 1 Coronary angiography showing anteroposterior cranial view of case 1.

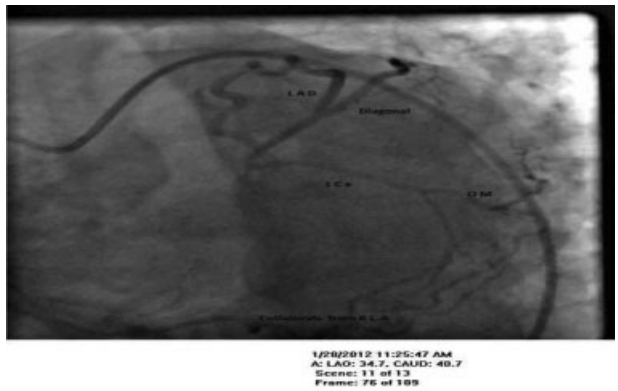

Fig. 2 CAG showing LAO caudal view of case 1. CAG, coronary angiography; LAO, left anterior oblique.

Second case was a middle aged (48 years) male, diabetic, hypertensive, current smoker, morbidly obese patient (having a body weight of 95 kg and height of 1.72 m, body mass index of 32.11 kg/m^2) presented with acute anginal chest pain, diaphoresis, and shortness of breath. ECG at the time of admission showed ST elevation, inferior wall myocardial infarction with ventricular tachycardia, and without any hemodynamic instability. In view of absence of contraindications and after obtaining informed consent, thrombolytic therapy was given with streptokinase (1.5 million units in 100 mL normal saline over 45 minutes). At presentation the BP was 100/70 mm Hg and heart rate was 150 per minute. After thrombolytic therapy the clinical symptoms, ECG changes and ventricular arrhythmia subsided. It was accompanied by standard treatment with dual antiplatelet agent (aspirin 150 mg, and clopidogrel 75 mg once daily), β blocker (metoprolol succinate), ACE inhibitor (ramipril), analgesics and sedatives (intravenous morphine 3 mg). The BP improved to 140/90 mm Hg, heart rate became regular

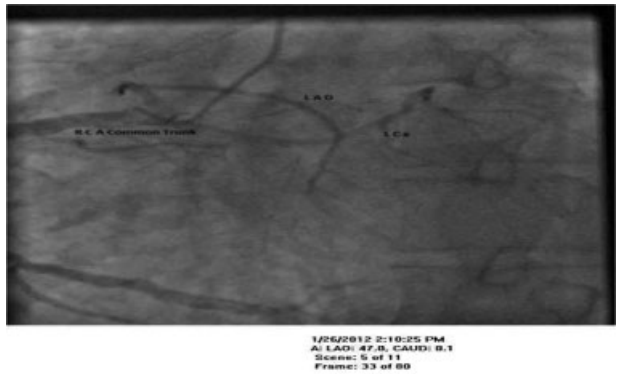

Fig. 3 CAG showing LAO view of case 2. CAG, coronary angiography; LAO, left anterior oblique.

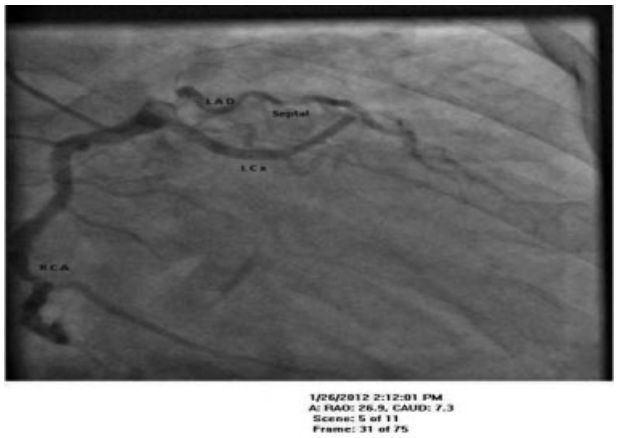

Fig. 4 CAG showing RAO view of case 2. CAG, coronary angiography; RAO, right anterior oblique.

with 80 beats per minute. Echocardiogram of the heart revealed regional wall motion abnormality of the inferior and posterior wall with mild left ventricular systolic dysfunction (EF = 48%). The cardiac biomarker troponin-I was elevated (10.2 ng/mL). The patient's LDL, HDL and total cholesterol were found to be 180, 28 and 290 mg/dL, respectively. The CAG revealed a single coronary artery arising from right aortic sinus. From proximal part of this right common trunk both anterior descending and circumflex branch were originating and supplying the left heart with combined diverse course, as shown in ►**Fig. 3** and ►**Fig. 4**. The right coronary artery showed diffuse atheromatous disease without significant stenosis in major branch. This is of R-III C type single coronary anomaly as per Lipton classification. CAG was completed through right radial route. As there was no occlusive stenosis in any major branch, the patient was given optimal medical management using dual antiplatelet (aspirin 150 mg, and clopidogrel 75 mg once daily), β blocker (metoprolol succinate), ACE inhibitor (ramipril), statin (atorvastatin 80 mg) and oral antidiabetic agent on discharge. The case also underwent multi-slice computerized tomography (MSCT) CAG and treadmill test during follow-up.

Discussion

Normally, coronary arteries have two or three coronary ostia. Two ostia (right and left) are typically present. The third coronary artery or conal or infundibular branch is present in 23 to 51% of normal hearts. The exact pathogenic mechanisms for development of single coronary artery syndrome are not known. They are usually congenital malformation of a normal coronary system that is dependent on multiple morphologic features, including formation of cardiac sinusoids, development of coronary buds on embryologic aortopulmonary trunk and selective connection between the two systems. The anomaly most associated with sudden death is a left main coronary artery arising from the right aortic sinus, and traverses between two great arteries (Lipton RII B or LII B). The high risk features are slit-like ostium, acute angle take off, and interarterial course. Usually the diagnosis is made by conventional CAG, MSCT-CAG and cardiovascular magnetic resonance angiography. Advantages of MSCT-CAG include detailed information about coronary artery course and disease, detailed structural information preoperatively as guidance for corrective procedures or surgeries, and short acquisition time for images. It can effectively disclose high-risk features for sudden cardiac death in this anomaly such as: slit like ostium, acute angle take off, interarterial course, and demonstrates crucial advantage over invasive CAG.[10] Prognosis for patients with a single coronary artery varies from excellent with no decrease in life expectancy[11,12] to sudden death.[13]

Conclusion

The key importance of single coronary artery resides in difficulties of diagnosis during CAG and cardiac surgery. Every coronary angiographer should, therefore, be familiar with the existence and anatomical types of this congenital anomaly. During cardiac surgery unexpected complications may arise. In the light of the rarity of this condition, it seems unlikely that the pathological importance of a single coronary artery in the absence of coronary atheromatosis will ever be completely elucidated. As a consequence, therapeutic strategies will mostly remain speculative.

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